keragel®
& kerageI®T
In the Management of Epidermolysis Bullosa

This booklet is aimed at helping people who have Epidermolysis Bullosa (EB), families, carers and health professionals. The Standard Therapies and approaches to wound management are listed on page 2. This booklet seeks to ensure that the best possible treatment outcomes are achieved for patients when using keragel®/kerageI® products in combination with these Standard Therapies and the International Consensus Best Practice Guidelines for Skin and Wound Care in Epidermolysis Bullosa.

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Standard Therapies for EB

By definition, inherited EB is a genetically transmitted disorder characterized by marked fragility of the skin. Any trauma, no matter how minimal it may seem, is likely to cause the skin of an EB child or adult to tear or blister. The following are recommended ways to avoid or minimize this problem:

1. **Reducing friction...**
   Extreme care should be employed in handling the skin of any patient with EB.

2. **Non-adhesive bandages and dressings...**
   Adhesive or semi-adhesive dressings, bandages, Band-aids or tape should not be used on the surface of the skin. Instead, wounds should be covered with an appropriate non-adhesive dressing and then further wrapped loosely with rolled gauze. This can be secured by using a tubular dressing retainer.

3. **Keeping the skin cool...**
   Nothing hot should ever be applied to the skin of a patient with EB. In particular, bath water should be no warmer than body temperature. Patients should avoid prolonged exposure to ambient heat and humidity. If possible, air conditioned environments should be sought whenever possible.

4. **Managing blisters...**
   Because blisters in EB are not self-limiting, and can fill with fluid and grow quite large, they need to be drained.

5. **Clothing...**
   In younger children, diapers may require additional padding at the legs and waist. Whenever possible, loose-fitting garments should be worn. If blisters develop from the seams of clothing, garments may be worn inside-out and tags, cuffs and necklines may be removed. Loosely-fitted, padded shoes are generally better tolerated.

6. **Nutritional deficiencies...**
   Many children with EB become anaemic due to a chronic loss of blood through wounds, poor nutritional intake, poor absorption of iron and bone marrow suppression from chronic inflammation. It is important to work with a nutritionist experienced in the care of special needs patients. Treatment for iron deficiency anaemia is often necessary. Other patients have selenium and carnitine or vitamin D deficiencies which may predispose them to cardiomyopathy and osteoporosis. Many patients develop failure to thrive and require feeding gastrostomies.

7. **Monitoring for cancer...**
   Squamous cell carcinoma is the leading cause of death in EB usually occurring after the 2nd decade of life. Patients with RDEB and JEB are at increased risk of developing skin cancers during their lifetimes. It is very important that all EB patients have at least yearly examination of all skin areas.

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http://rarediseases.org/rare-diseases/epidermolysis-bullosa/
What are keragel® & keragelIT®?

keragel® & keragelIT® are keratin-rich hydrogels. keragel® is a thicker gel which has superior exude handling. keragelIT® is a thinner gel which is faster drying. They are made in New Zealand and the keratin is extracted from sheep wool.

What is keratin?

Keratin is a class of protein. Keratin proteins are one of the dominant protein types in the skin in both the epidermis (first) and the dermis (second) layers.

Why does keratin help heal wounds?

Keratin proteins have been shown (in laboratory tests) to be able to signal to cells in the skin (keratinocytes and fibroblasts) in a way that affects their migration and proliferation. This is needed for wound healing. By inoculating cell media with keratin proteins, it has been shown² that the cells enter a hyper-proliferative phase where migration rates are increased and the expression of key wound healing proteins (such as Collagen IV & VII) is also increased. This means that the keratin may be able to help the cells in the wound to proliferate and migrate more quickly and the wound may epithelialise faster. Further, the healed wound may have more of the key proteins that are needed to strengthen the skin. This idea is illustrated in the diagram on the next page.
Why is keratin-assisted wound healing beneficial for EB?

Accelerated healing of wounds is particularly beneficial to people with EB since wounds occur so frequently. The genetic defect typically means that people with EB have insufficient or dysfunctional structural proteins, which makes the skin weak. If the changes to the protein expression of keratinocyte cells observed in-vitro occur in-vivo then applying the keratin topically to the wound may be able to elevate the number of structural proteins made and thus strengthen the skin. Improvements in wound management for people with EB have been seen in studies conducted\(^3,4,5\).
General recommendations for using keragel® & kerageIT®

PREPARATIONS:
1. Cleanse the wound in accordance with normal procedures.
2. Ensure that the skin surrounding the wound is dry.

3. The tube is opened using the piercer cap to break the nozzle seal. To ensure sterility, sanitise the piercer cap before using.

4. We recommend if using keragel®/kerageIT® on a large surface area of skin to dispense keragel®/kerageIT® into a sterile container.

5. The keragel®/kerageIT® will then be applied using sterile gauze (as shown to the side). This reduces risk of infection compared to using fingers or gloves.
When to use keragel® & when to use keragelIT® and how to apply?

keragelIT® dries more quickly than keragel® and thus is recommended when the wound is able to be left uncovered or covered with a tubular or crepe bandage. If the wound needs a secondary dressing for protection then either keragel® or keragelIT® can be used (whichever you or your health professional prefer), this will be described in more detail on page 8.

Applying keragelIT® on wounds which don’t require any other dressings

Typically, people with EB find it most comfortable to use keragelIT® alone (no other covering dressing) for wounds on the face and sometimes on the neck, hands and feet. To do this, apply a thin layer of keragelIT® by dabbing it on with sterile gauze then allow the keragelIT® to dry. Approximately 30 seconds after application, it will become sticky/tacky and after approximately 5 minutes it will dry to a shiny layer and when bandages/socks etc. can be put on top if required. A dusting of cornflour can be used after it has become tacky to help it to dry a little faster.

This person found keragelIT® alone to be most comfortable on her neck. She applied it daily and allowed it to dry before putting her clothes back on. The coating of keragelIT® keeps the nerve endings moist, which aids healing and also keeps the air off the wound, which can help with pain that this might normally cause. It can also reduce itch and hence the damage that would occur from scratching.

If you are washing these areas daily, then you will need to reapply keragelIT® thereafter as it is water soluble.
Applying kerageIT® on wounds which require simple protective dressings

Often people with EB like the additional protection offered by a dressing or tubular bandage placed over the top of kerageIT®. In the case shown (photo to left) a thin layer of kerageIT® is being applied by dabbing it on with sterile gauze. Allow the kerageIT® to dry. Approximately 30 seconds after application, it will become sticky/tacky. Tubular bandages/socks etc. should not be put on immediately.

After approximately 5 minutes it will dry to a shiny layer and now bandages/socks etc. can be put over it. A dusting of cornflour or some bland moisturising cream can be used after it has become tacky to help it to dry it a little faster so you don’t have to wait as long to put bandages etc. over it.

In this case (photo to right), kerageIT® has been allowed to become tacky and then moisturising cream has been applied to allow bandages to be applied without waiting for it to dry fully.

The completely ABC-wrapped fingers can be seen below.

For wounds on the feet (photo below), using kerageIT® means just a short wait before socks can be put on.
Applying keragel®/keragelT® on wounds which require protective dressings

On areas which need additional protection, people who have EB often choose to use a protective dressing. In this case, the faster drying nature of keragelT® is not essential and either keragel® or keragelT® (whichever you or your health professional prefer) can be used.

The keragel®/keragelT® should be dispensed as shown on page 5 and then applied to the secondary dressing with sterile gauze as shown in the picture on the left. A layer 1−2 mm (approx. 1/16”) thick should be used.

The dressing with the keragel®/keragelT® on it should then be placed on the wound as in the picture on the left.

If you are using a thicker type absorbent dressing for exudate management or extra protection, then it is recommended that keragel® is used. This is a thicker gel and won’t be excessively drawn into the dressing which may reduce its effect.
What secondary dressings are recommended for use with keragel®/keragelIT®?

Atraumatic (non-adhesive) dressings that won’t disturb the wound when removed are recommended for use as protection over keragel®/keragelIT®. Dressings that stick and cause trauma when removed should be avoided. If a dressing has stuck then add water to help remove it (keragel®/keragelIT® are water soluble), alternatively use a medical adhesive remover (as recommended by your health professional). Superabsorbent dressings that will strongly extract the keragel®/keragelIT® out of the wound are not recommended.

Recommended care

The wound should be inspected every 2-3 days or following the established dressing regime as recommended by your health professional. If the wound is looking dry, then more keragel®/keragelIT® should be applied to the wound area. Clean the wound first and then apply new keragel®/keragelIT®. For wounds with no covering dressing, apply keragelIT® at least daily since it gets washed off. It is recommended that application of keragelIT® is continued after a wound has healed to assist with skin remodelling phase and that further keragelIT® is used on skin surrounding the wounds and on skin considered to be vulnerable to wounding.

Additional tips for Managing stinging

keragel®/keragelIT® can be painful if applied to fresh wounds. The following are suggested ways to minimise and manage stinging:

1. Apply the keragel®/keragelIT® to a small wound that is not too ‘raw’. If there is no stinging then progressively try larger wounds or wounds that are more ‘raw’. If it does stings try one of the methods below. If these do not reduce stinging to an acceptable level then try again after a few days.

2. Apply the keragel®/keragelIT® to a non-adherent dressing and then place the coated dressing on to the wound. (as shown on page 8).

3. Dilute the keragelIT® with sterile water at a 2:1 ratio before application, e.g. 10gms keragelIT® to 5 mls water.

4. Mix keragel® with a bland moisturiser or emollient as suggested by your health professional.

5. Warm the keragelIT® to body temperature, e.g. immerse the tube in 40°C water before use.

6. For new blisters, if it is too painful to apply keragelIT® after lancing, keragelIT® can be applied to the roof of the blister before it is lanced.
Use of keragel® & kerageIT® on new blisters

Best practice guidelines suggest that new blisters (such as the one above) are lanced & drained either with a sterile blade or needle, as in the pictures to the right.

It is recommended that keragel®/kerageIT® is applied to the roof of such blisters, after they are lanced. However, if this causes too much stinging, keragel®/kerageIT® can be applied before lancing - refer Tip 6 on Pg 9. If the blister roof is absent then check that keragel®/kerageIT® can be tolerated if applied directly on to the wound bed.

Infected wounds?

If the wound is clinically infected then an antimicrobial product should be used to manage the infection. keragel®/kerageIT® do not have antimicrobial properties but they may be used in combination (mixed together 50:50) with antimicrobial dressings such as PHMB, Flaminal®, silver or honey. Alternatively, the use of keragel®/kerageIT® may be suspended until the infection is cleared and then reinstated.
Odour

keragel® and kerageIT® have a distinctive odour - this is the smell of keratin protein. Some people like it, some people don't. The smell is strongest when the gel is freshly squeezed from the tube. The smell is barely noticeable once the gel is dry on a wound and impossible to smell under other dressings.

What to expect from keragel®/kerageIT®?

For acute wounds (less than 6 weeks old), you can expect to see accelerated healing at each dressing change. If an acute wound is failing to respond (e.g. reduce in size) in 14 days then consult your Health Professional.

For chronic wounds (more than 6 weeks old), you can expect to see some healing (e.g. reduction in size), but relative to the time it has been there. If you are treating a wound that is 10 years old, then it may take several months to heal. If a chronic wound is failing to respond (e.g. reduce in size) in 2 months then consult your Health Professional.
REFERENCES


ADDITIONAL INFORMATION

1. www.keraplast.com/epidermolysis-bullosa

2. The Keraplast Wound Care YouTube channel at www.youtube.com

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